Acute rheumatic fever in Hawaii: 1966 to 1988

L T Chun MD
D Venu Reddy MD MPH
Gregory K Yim MD
Loren G Yamamoto MD MPH

Seventy five children with acute rheumatic fever (ARF) were hospitalized on Oahu from 1984 to 1988. The annual incidence rate was 9.5 (all rates are per 100,000 children per year). The first attack and recurrent attack rates were 7.9 and 1.6. Polyarthritis occurred in 84%, chorea in 7%, and carditis in 32%. Mitral insufficiency was the most common valvular lesion (88%). Hawaiians/part Hawaiians and Samoans had the highest incidence rates (relative risk 3 and 56, respectively). Polynesian children were 84 times more likely to develop carditis.

Five hundred thirty nine ARF cases were hospitalized on Oahu, 1966 to 1974 and from 1976 to 1988. The annual incidence rate of ARF on Oahu has remained fairly constant at about 12.4. The incidence rates in all ethnic groups have decreased except in Samoan children.

Introduction

In the early 1980s, numerous articles appeared that relegated ARF to a minor role in the continental United States among children in the 5 to 17-year age group[1,2,3]. As compared to 40 to 65 cases per 100,000 population per year between 1935 and 1960, the number of cases declined to 0.5 to 1.9 cases per 100,000 population per year between 1977 and 1981[4,5]. Clinicians in Hawaii, however, have continued regularly to encounter patients with ARF. A resurgence of ARF in the continental U.S. occurred in 1985, the attack rate increasing to 18 per 100,000 population per year in Salt Lake City, Utah[6,7]. Since 1985, others have reported similar experiences[6-15]. Because of the renewed interest in ARF, we were prompted to continue our survey of hospitalized children with ARF in Hawaii from 1984 to 1988. We previously reported our experience with ARF for the periods 1976 to 1980[16] and 1980 to 1984[17] in which the annual attack rates were 14.4 and 13.4 per 100,000 children respectively.

Historically, the first case of ARF in Hawaii was described in a medical conference in 1938[18] and the first recorded case was reported in 1941[19]. During 1942 to 1946, 17% of all cardiac cases admitted to the hospitals (1.2% of all admissions) in Hawaii were due to rheumatic fever (RF)18. Between 1947 and 1949, 73 patients (1.2% of all admissions) were admitted for RF[21]. In 1950, the Hawaii Territorial Board of Health had on its active clinic roster 257 acute and chronic pediatric RF cases. Between 1955 and 1959, 91 children (0.6%) with RF were admitted to the Kauikeolani Children's Hospital in Honolulu[20]. Children hospitalized in the 7 major hospitals on Oahu, with first attack ARF from 1966 to 1974, accounted for an annual rate of 11.8 cases per 100,000 children aged 0 to 19 years[22].

Hawaii differs from the continental U.S. in that it is geographically isolated and its ethnic composition is composed largely of Polynesians, Asians, and Caucasians. Epidemiological patterns of diseases other than RF often resemble the patterns in North America and Asia reflecting Hawaii's geographical location between those two land masses, as well as Hawaii's genetic blend of people from both the west and the east sides of the Pacific ocean.

Hawaii lends itself to the study of epidemiology because its borders are clearly defined, making it easier to perform population-based surveys of the occurrence of disease. RF in Hawaii has been regularly studied since the 1940s[16-23]. The purpose of our report is to describe the most recent survey of ARF from 1984 to 1988 and to present our overall experience with ARF over a 20 year period from 1966 to 1974 and 1976 to 1988.

Methods

The case records of children 4 through 18 years of age who were hospitalized October 1984 through September 1988 for ARF in all 10 hospitals on the island of Oahu (City and County of Honolulu) were reviewed. The diagnosis of ARF was based on the revised Jones criteria[24]. Non-hospitalized children and those not residing on Oahu were not included in this study.

The Hawaii State Department of Health Research and Statistics Office provided 1986 estimates of the population of Oahu children 4 through 18 years of age. This was divided into 8 major ethnic groups: Caucasian, Japanese, Chinese, Filipino, Hawaiian/part-Hawaiian, Samoan, Black and others. Samoans were defined as children with 100% Samoan ancestry. Tongan children, children with mixed ethnicity, other Pacific island children were included in the "other" category. This group was composed mostly of those with mixed ethnicity. Due to the heterogenous ethnicity in Hawaii, this group did not have any single dominant ethnic population. Hawaiians and part-Hawaiians were grouped together.

The data collected from 1984 to 1988 were compared with similar data collected in 3 previous studies done on Oahu which examined groups studied in 1966 to 1974[22], 1976 to 1980[16], and 1980 to 1984[17]. Although the latter 2 studies included children with first attack as well as recurrent ARF, the 1966 to 1974 study examined children only with first attack ARF.

Results: 1984 to 1988

Seventy-five children with ARF were identified in the study hospitals during the 4-year period. In each of those 12-month

Department of Pediatrics
University of Hawaii John A Burns School of Medicine
Kapiolani Medical Center for Women and Children

Address reprint requests to: D Venu Reddy MD MPH
1319 Punahou St, Suite 1160, Honolulu HI 96826

Table 1. Age and Sex Distribution of ARF Patients.

GROUPS	10/84-/88	10/80-9/84	10/76-9/80	OVERALL
4 -5 years	2 (3%)	4 (4%)	5 (5%)	114 (4%)
6 - 11 years	38 (51%)	43 (44%)	52 (50%)	133 (48%)
12 - 15 years	2 (39%)	34 (35%)	35 (34%)	98 (35%)
16 -18 years	6 (8%)	17 (17%)	12 (12%)	35 (13%)
Males	4 (65%)	59 (60%)	53 (51%)	161 (58%)
Females	26 (35%)	39 (40%)	51 (49%)	116 (42%)
Total	75	98	104	277

Table 2. Month of onset of ARF

MONTHS	10/84-9/88	10/80-9/84	10/76-9/80	OVERALL
January	4 (5%)	7 (7%)	13 (13%)	24
February	5 (7%)	8 (8%)	8 (8%)	21
March	2 (3%)	4 (4%)	8 (8%)	14
April	8 (11%)	14 (14%)	6 (6%)	25
May	6 (8%)	3 (3%)	2 (2%)	11
June	4 (5%)	6 (6%)	2 (2%)	12
July	2 (3%)	4 (4%)	5 (5%)	11
August	8 (11%)	10 (10%)	13 (13%)	31
September	12 (16 %)	16 (16%)	7 (7%)	35
October	9 (12%)	9 (9%)	17 (16%)	35
November	6 (8%)	10 (10%)	9 (9%)	25
December	9 (12%)	7 (7%)	14 (13%)	30

periods there were respectively, 10, 11, 24, and 30 hospitalized children with ARF. The 1986 Oahu population for the age groups from 4 through 18 years of age was 197,174, so that the average annual incidence was 9.5 per 100,000 children. The annual incidence for first attacks was 7.9 per 100,000 children (62 cases) and that for recurrent acute attacks was 1.6 per 100,000 children (13 cases). At the time of the recurrent acute attack, none of the children had been on antimicrobial prophylaxis

The age and sex distribution is shown in Table 1. The majority of the cases occurred in the 6 through 15 year-old age group. Sixty-five percent were boys and 35% were girls. The monthly distribution in the 1984 to 1988 study period and during the study period 1976 to 1984 is shown in Table 2. Most of these cases occurred between the months of August and December.

The overall incidence of RF and carditis is broken down by ethnicity in Table 3. Hawaiians/part-Hawaiians and Samoans (Polynesians) had the highest incidence, accounting for 72% of the cases. As compared to Caucasian children, Hawaiian/part-Hawaiian children were 3 times and the Samoan children were 56 times more likely to have ARF. There were no Blacks with ARF during the study period.

Carditis occurred in 24 children (32%). Although the numbers are small, one can say that the Polynesian children were 84 times (relative risk 84 compared to the incidence rate of non-Polynesians) more likely to develop carditis. Of the children with carditis, 2 had Sydenham's chorea and 8 had recurrent attacks of ARF. Mitral insufficiency (MI) occurred in 21 children (88%) (13 children had MI alone). Combined mitral and aortic insufficiency (AI) occurred in 8 children (33%), and isolated AI occurred in 2 children (8%). Congestive heart failure was present in 10 children (42%). Pericarditis occurred in 4 children (17%). One child had third-degree atrioventricular block (AVB) along with MI, AI, and congestive heart failure.

Table 3. Rheumatic fever and carditis in children 4 through 18 years of age on Oahu, Hawaii, by Ethnicity, 1984 to 1988. Population figures are for nonmilitary population.

Ethnic Groups	Population		RHEUTMATIC FEVER			CARDITIS	
	Number	Pct	# of Cases	Incidence*	Relative Risk**	# of Cases	Incidence
Caucasian	36,125	18.3	5	3.5	1.0	0	0
Japanese	28,829	14.6	0	0	0	0	0
Chinese	7,062	3.6	0	0	0	0	0
Filipino	21,813	11.1	3	3.4	1.0	0	0
Hawaiian/ part Hawaiian	49,113	24.9	23	11.7	3.4	6	3.1
Samoan	3,981	2.0	31	195.0	56	13	82
Black	9,067	4.6	0	0	0	0	0
Other	41,184	20.9	13	7.9	2.3	5	3.0
Total	197,174		75	9.5		24	3.0

* Incidence is the rate per 100,000 population per year.
** Relative risk uses the Caucasion group as the denominator.

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Of the other major manifestations, polyarthritis occurred in 63 cases (84%), chorea occurred in 5 (7%) and erythema marginatum occurred in 2. There were no cases with subcutaneous nodules during this period. Polyarthritis and carditis occurred together in 14 cases. Polyarthritis and chorea occurred together in 1 case. As stated above, chorea and carditis occurred together in 2 cases.

First degree AVB was present in 22 out of 69 children based on the electrocardiogram (32%). Fever was present in 41 children (55%). Arthralgia was reported in 50 children (67%). Arthralgia was noted to be polyarthritic; additional minor criteria were used to fulfill the Jones criteria in these instances.

[It is to be noted that 32 of the 75 children (43%) had no medical insurance and 3 (4%) had Medicaid coverage. Of the 35 patients on Medicaid, or who had no medical insurance, 29 (83%) were Polynesian. Polynesians without insurance had a higher incidence of ARF than the Polynesians with insurance.]

Results: 1966 to 1988

During the 20-year period 1966 to 1974 and 1976 to 1988, 539 hospitalized cases of ARF in children were observed on Oahu. Although the total number of new cases of ARF is declining on Oahu, the incidence pattern differs from that of the continental U S.

The number of ARF cases identified during 1966 to 1974 was 262, which is approximately 29 cases per year[22]. A similar trend continued in 1976 to 1980 (104 cases) and 1980 to 1984 (98 cases) when approximately 26 and 25 cases respectively occurred annually[16,17]. Our survey, on the other hand, in 1984 to 1988, showed a decreased number of cases, a total of 75, which is 19 cases per year. Fig. 1 describes the number of ARF cases over the 20-year study period among the different ethnic groups.

During the period from 1966 to 1974, only children with first attack ARF were considered. The annual incidence was 11.8 hospitalized cases per 100,000 children[22]. During the 1976 to 1980[16] and 1980 to 1984[17] periods, the first attack rates were 12.5 and 10.9 hospitalized cases per 100,000 children, respectively. Samoans were not categorized separately in the 1966 to 1974 period. They were included in the "other" group during this period.

The overall annual incidence of ARF for both first-attack and recurrent acute attacks was 14.4 and 13.4 hospitalized cases per 100,000 children during the 1976 to 1980[16] and 1980 to 1984[17] periods, respectively. Fig. 2 shows the incidence among the ethnic groups during each of the two study periods covering 1976 to 1988. Samoan children had an average annual incidence of 166 per 100,000 and Hawaiian/part-Hawaiian children had an average annual incidence of 19 per 100,000. This compares well with Caucasian children who had an annual incidence of 5 per 100,000. The incidence trends show a low baseline rate for Japanese and Chinese children, a low declining rate for Caucasian and Filipino children, a higher but declining rate for Hawaiian/part-Hawaiian children, and a persistently high rate in Samoan children.

Discussion

The incidence of rheumatic fever had been declining in the continental United States. Articles describing this decline appeared in literature as late as 1985[1,2,3]. More recent reports have described a sudden resurgence of rheumatic fever[6-10,12-15].

The reasons for the dramatic decline in the incidence of acute rheumatic fever in the United States and other highly developed countries have never been clearly established[25]. The associations between it and poverty, living standards, socioeconomic status, nutrition, hygiene, access to medical care, antibiotics, degree of crowding in households and genetics have all been implicated. Another factor contributing to first the decline and then to its resurgence has been the changes in the biologic properties and clinical virulence of prevalent streptococcal strains, particularly M-serotypes (types 1, 3, 5, 6, 14, 18, 19, 24) which have been implicated as markers of rheumatogenicity. These are highly virulent and strongly associated with ARF[25].

The incidence of RF in Honolulu was constant during 1966 to 1984 and then declined during 1984 to 1986, with a subsequent rise in 1987 and 1988. The high percentage of Asian and Polynesian children in Hawaii makes the ethnic composition of our population very different from that in other states. However, in observing the trend in the Caucasian group alone, a significant rise in incidence among this group has not been observed. Besides that, the trends in the Asian and Hawaiian/part-Hawaiian groups have been consistently downward.

It is clear that our data indicate that the incidence in Hawaiians/part-Hawaiians and in Samoans is many times higher than in other ethnic groups. Although the absolute incidence and the incidence in Hawaiian/part-Hawaiians are both declining, the reason for the opposite trend for the incidence of Samoans is unclear. The rate is determined by comparing the absolute incidence with the total Samoan population of this age group. The absolute incidence among Samoans appears to be roughly the same in 1980 to 1988.

The reasons for these ethnic differences have been discussed in previous reports[16,17]. Newer data attempting to further clarify this issue have not been forthcoming, as yet.

Utah described the ethnic breakdown of its RF patients as 94% Caucasian, 4% Pacific Islander, 1% Hispanic, and 1% Asian; this was similar to the percentages of its general population[6,7]. The Pennsylvania cohort of RF patients during 1985 to 1986 was 94% Caucasian; this was an increase from 81% Caucasian in the 1965 to 1974 period. The frequency of Blacks in this cohort decreased from 15% to 0% during the same time period[9]. The cohort of RF patients in Columbus, Ohio, was 90% Caucasian and 10% Black in a community composed of 80% Caucasians and 20% Blacks[8]. The RF cohort reported from Akron, Ohio, was 96% Caucasian[13]. These recent series of RF patients suggest that Caucasians are more frequently affected than other ethnic groups. (This trend has not been observed on Oahu.)

A series of 110 RF patients seen over a period of 20 years in New York City noted a shift in ethnic distribution with an increase in Hispanics and a decrease in Blacks which paralleled the ethnic distribution of the population seen in their outpatient clinic[12]. The series of cases in Tennessee had demographic characteristics similar to the demographics of the population in general served by the study center, which included an urban, low-income group of large families[14]. These ethnic comparisons do not indicate a consistent pattern across the country as a whole.

In a national survey of medical centers across the U.S. mainland, 5,019 cases of RF were identified during the 4-year period from 1984 to 1987. More than 50% had heart involvement.

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By examining the change in incidence of RF during this period in different regions of the U.S. investigators were unable to demonstrate any significant nationwide trend in RF. The survey suggested regional increases which were not present in all areas of the U.S.[11].

The epidemiology of RF on Oahu appears to have an incidence pattern of its own. These data add support to the conclusion that recent increases in RF are regional rather than universal.

Cases of rheumatic fever occurred less commonly during the months of May, June, and July in our series. Utah noted a peak incidence in March and April[6]. The series in Columbus noted a higher frequency in March through August[8].

Carditis occurred in 78% and chorea occurred in 28% of the Utah cohort. This cohort also noted carditis and polyarthritis together in 44%, carditis and chorea together in 14%, and carditis, chorea, and polyarthritis together in 6%[6]. In the Pennsylvania cohort, carditis occurred in 59%, carditis and polyarthritis in 45%, and carditis and chorea in 17%[9]. In the Columbus cohort 50% had carditis, 18% had chorea, and 65% had polyarthritis[8]. Carditis occurred in 30% and chorea occurred in 9% of the Akron cohort[13]. The series from Tennessee reported carditis in 75% and chorea in 29%[14]. The New York cohort noted 51% with carditis and 4% with chorea[12]. The low frequency of chorea in the New York series was attributed to the practice of some neurologists to follow chorea in an outpatient setting[12].

The frequency of chorea and carditis in most of the Mainland cohorts is much higher than that observed in our Oahu cohorts. It is possible that the frequency of carditis may be underestimated in our cohort during this time period because of the limited use of Doppler echocardiography. However, it is unlikely that this alone could account for such a large difference. It is unlikely that the frequency of chorea could be underestimated, because it is quite a dramatic presentation. An alternative explanation might be that in other communities, hospitalization of RF patients with polyarthritis alone is not a standard practice. Bedrest and salicylate therapy can reasonably be administered to an outpatient; of 23 patients with acute RF during 1986 in the Akron series, only 14 were hospitalized[13].

If treatment of these RF patients is done as an outpatient, then studies examining the frequency of RF among hospitalized cohorts will not identify these patients. Patients with polyarthritis alone are less likely to warrant hospitalization than patients with carditis and chorea. On Oahu in general, all children with RF are hospitalized, even if polyarthritis is the only criterion apparent.

It is possible that in the above-mentioned Mainland series, researchers have failed to identify a significant portion of children with RF presenting with polyarthritis alone. Outpatient treatment with salicylates or nonsteroidal anti-inflammatory drugs for what might have been identified as joint pain due to trauma or other forms of reactive arthritis may have prevented the identification of these patients as having of RF that required hospitalization. The period of time during which RF was almost nonexistent in many areas may have been the result of failure on the part of clinicians to consider this diagnosis in a patient with joint pain alone. Among pediatricians in Honolulu, this is less likely to occur since RF has never really disappeared from Hawaii.

One recent article described the identification of carditis by means of Doppler echocardiography in patients who clinically appeared to have chorea without carditis[7]. Echocardiograms were done on 36 patients in our cohort. Echocardiograms identified carditis in 17 cases. Seven patients with carditis did not have echocardiograms done. Of the 5 patients with chorea, 2 had echocardiograms done; one was negative and one had identifiable carditis. We did not record whether the diagnosis of carditis had been identified clinically prior to the echocardiogram.

On examining the trend of carditis from 1976 to 1988, the frequency of carditis continued to decrease. Echocardiography was performed only infrequently prior to 1984 in our cohort. This suggests that the routine use of echocardiography has not significantly increased diagnostic identification of carditis in RF patients in this cohort. However, in other cohorts where the frequency of carditis is higher, routine Doppler echocardiograms may be of value in identifying occult valvulitis.

Seventeen percent of the 1984 to 1988 Oahu RF cases were due to recurrent RF. The frequency of recurrent cases was 19% in the 1980 to 1984 series[17] and 13% in the 1976 to 1980 series[16] on Oahu. None of the patients in the 1984 to 1988 and the 1980 to 1984 Oahu RF series with recurrent RF was on antibiotic prophylaxis at the time of recurrence[16,17]. A 10% recurrence rate was noted in the New York series. Ten of those 11 recurrences took place in patients who were not on antibiotic prophylaxis at the time of recurrence[12]. This suggests that recurrence of RF can be effectively prevented if compliance with antibiotic prophylaxis is continued.

A program established by the Hawaii State Department of Health using public health nurses, clinic nurses, and pediatric cardiologists to follow children with RF until they become adults helps to enforce compliance. Despite this program, the frequency of recurrence is still significant, although only among patients not on antibiotic prophylaxis. The island community of Oahu has clearly defined borders and has a relatively less mobile population. However, in other regions where similar programs to enforce compliance with antibiotic prophylaxis exist, keeping track of RF patients would be significantly more difficult, considering that there are no barriers there to unrestricted mobility.

Summary

The total incidence of RF on Oahu has remained fairly constant. The incidence among all ethnic groups has decreased, with the exception of the Samoans. Polynesians have the highest incidence in all of the studies examining RF in Hawaii. The ethnic changes and incidence trends of RF in areas of the U.S. mainland described in the literature have not been duplicated in Hawaii. The lower frequency of carditis and chorea among RF patients on Oahu is not easily explained. Despite its nearly vanishing from areas of the Mainland in the recent past, RF has never disappeared from Hawaii. Theories attempting to account for the fluctuation in the frequency of RF must also explain the lack of such fluctuation in Hawaii, in order to be valid.

REFERENCES

1. Land MA, Bisno AL. Acute rheumatic fever: A vanishing disease in suburbia. *JAMA*. 1983;249:895-898.
2. Markowitz M. The decline of rheumatic fever: Role of medical intervention. *J Pediatr*. 1985;106:545-550.

3. Gordis L. The virtual disappearance of rheumatic fever in the United States: Lessons in the rise and fall of disease. *Circulation.* 1985;72:1155-1162.
4. Holmberg SD, Faich GA. Streptococcal pharyngitis and acute rheumatic fever in Rhode Island. *JAMA.* 1983;250:2307-2312.
5. Schwartz RH, Hepner SI, Ziai M. Incidence of acute rheumatic fever: a suburban community hospital experience during the 1970s. *Clin Pediatr.* 1983;22:798-801.
6. Acute rheumatic fever: Utah. *MMWR.* 1987;36:108-110,115.
7. Veasy LG, Wiedmeier SE, Orsmond GS, et al. Resurgence of acute rheumatic fever in the intermountain area of the United States. *N Engl J Med.* 1987;316:421-427.
8. Hosier DM, Craenen JM, Teske DW, et al. Resurgence of acute rheumatic fever. *Am J Dis Child.* 1987;141:730-733.
9. Wald ER, Dashefsky B, Feidt C, et al. Acute rheumatic fever in western Pennsylvania and the tristate area. *Pediatrics.* 1987;80:371-374.
10. Ferrieri P. Acute rheumatic fever, the comeback of a disappearing disease. *Am J Dis Child.* 1987;141:725-727.
11. Kawasaki disease, rheumatic fever incidence assessed. *Infectious Dis in Child.* 1990;3(1):4.
12. Griffiths SP, Gersony WM. Acute rheumatic fever in New York City (1969 to 1988): a comparative study of two decades. *J Pediatr.* 1990;116:882-887.
13. Congeni B, Rizzo C, Congeni J, et al. Outbreak of acute rheumatic fever in northeast Ohio. *J. Pediatr.* 1987;111:176-179.
14. Westlake RM, Graham TP, Edwards KM. An outbreak of acute rheumatic fever in Tennessee. *Pediatr Infect Dis J.* 1990;9(2):97-100.
15. Giardina AV, Heaton S. Acute rheumatic fever in New York City. *NY State J Med.* 1988;88:385-386.
16. Chun LT, Reddy V, Rhoads GG. Occurrence and prevention of rheumatic fever among ethnic groups of Hawaii. *Am J Dis Child.* 1984;138:476-478.
17. Chun LT, Reddy DV, Yamamoto LG. Rheumatic fever in children and adolescents in Hawaii. *Pediatrics.* 1987;79:549-552.
18. Berk ME, Hartwell AS. Five years of heart disease in Hawaii. *Hawaii Med J.* 1949;8:177-180.
19. Doolittle SE, Tilden L. Rheumatic heart disease in Hawaii. *Hawaii Med J.* 1941;1:7-11.
20. Yim H. Rheumatic fever in Hawaii. *Am J Dis Child.* 1962;103:715-717.
21. Connor A, Yoshina T. Rheumatic fever in Hawaii. *Hawaii Med J.* 1951;10:181-182.
22. Rudoy R. Epidemiology of rheumatic fever in Hawaii, MPH thesis. University of Hawaii School of Public Health, Honolulu, Hawaii, 1979.
23. Reddy DV, Chun LT, Yamamoto LG. Advanced degree AV block in acute rheumatic fever. *Clin Pediatr.* 1989;28(7):326-328.
24. Committee on Rheumatic Fever and Bacterial Endocarditis (1982), American Heart Association: Jones Criteria (revised) for guidance in the diagnosis of rheumatic fever. *Circulation.* 1984;69:204A-208A.
25. Bisno AL. Group A streptococcal infections and rheumatic fever. *New Engl J Med* . 1991;325(11):783-793.

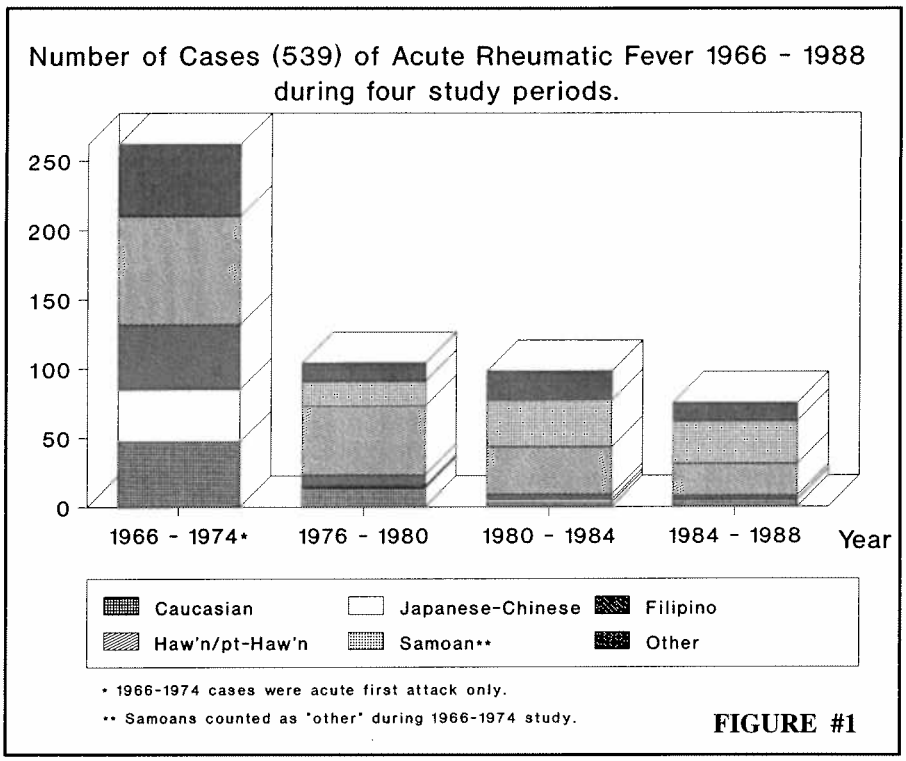

FIGURE #1

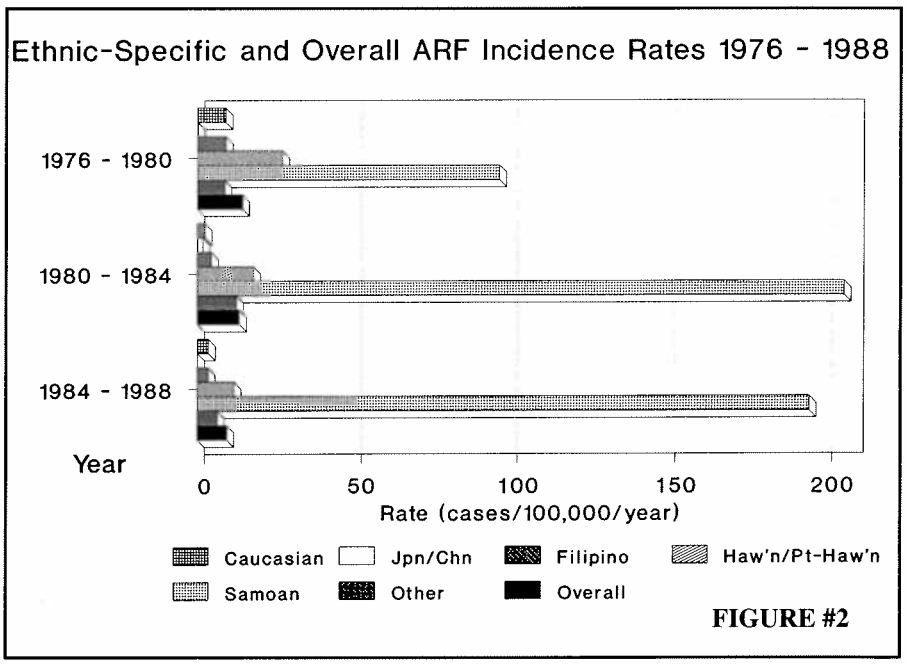

FIGURE #2